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CSF Pressure and CT Scan Findings in Cerebral Gigantism

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Cerebral gigantism is an unusual affliction marked by macrosomia, macrocephaly and almost universally mental retardation. In reviewing the clinical and laboratory evaluations of 5 children with this condition, it became apparent that this syndrome is markedly heterogeneous, indicating that it most likely arises due to a variety of different causes.

Using minimal diagnostic criteria that weight and head circumference should exceed the 95th percentile and that height should be near or above the 95th percentile (as originally described by Sotos), 5 children with this condition who have had extensive laboratory evaluations performed during the years 1974-81 were identified. Clinically, 3 were developmentally delayed or mentally retarded, and 2 were not. Routine laboratory analyses and endocrinologic studies were normal in all cases. When they were performed, EEG, skull xrays, audiometry, formal ophthalmologic examinations and inborn errors screens were normal. Head CT scans revealed heterogeneous results: one was normal, one demonstrated mild ventricular dilatation, one moderate ventricular dilatation, and two moderate dilatation of the subarachnoid spaces and ventricles. The latter two children, under chloral hydrate sedation, when fully asleep, had diagnostic CSF exams performed. The opening pressures were elevated in both, 296 and 230 mm H₂O, as were the closing pressures (200 and 150 respectively). Repeat CSF examination three months later in the child with the higher values revealed an opening pressure of 230 and closing of 190. The remainder of the CSF examinations were normal.

The clinical and laboratory heterogeneity in cerebral gigantism suggest that this condition may arise from different causes. The variable CT scan findings and elevated CSF pressures suggest that in some individuals communicating hydrocephalus may play a role. The decrease in pressure over three months suggests that this may be a transient phenomenon in the early childhood of some of these individuals.